

CLAIMS

SUB B1)

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1. A method of treating neurodegenerative disease in a mammal comprising the steps of introducing a therapeutic effective amount of a chaperone or chaperone-like-compound into the neurological system of the mammal.

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2. The method of claim 1, wherein the introducing step includes introducing the chaperone or chaperone-like-compound into the mammal by gene therapy.

SUB B2)

3. The method of claim 1, wherein the introducing step includes directly injecting the chaperone or chaperone-like-compound into the mammal.

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4. A method for screening for a test compound for chaperone-like activity for the treatment of neurodegenerative diseases comprising the steps of:

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introducing the test compound into transfected cells in tissue culture, wherein such transfected cells produce protein aggregate; and measuring the quantity of protein aggregate, wherein a test compound which decreases the quantity of protein aggregate as compared to control cells has chaperone activity.

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5. A method for screening for a test compound for chaperone-like activity for the treatment of neurodegenerative diseases comprising the steps of:

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introducing the test compound into an animal which models neurodegenerative disease;

allowing said animal to develop; and
subsequently measuring the quantity of
aggregates in said animal wherein decreased
aggregate formation over control animals indicates
chaperone-like activity.

6. A method of treating neurodegenerative
disease in a mammal comprising the step of
introducing a therapeutically effective amount of
a compound into said mammal wherein said compound
increases the effective concentration of a
chaperone in the neurological system.
7. A method of treating neurodegenerative
disease in a mammal comprising the step of
introducing a therapeutically effective amount of
a compound into said mammal wherein said compound
increases the effective concentration or enhances
the activity of a proteasome in the neurological
system.
8. A method for screening for a test compound
which increases proteasome activity for the
treatment of neurodegenerative diseases comprising
the steps of:
introducing the test compound into
transfected cells in tissue culture, wherein such
transfected cells produce protein aggregate; and
measuring the quantity of protein aggregate,
wherein a test compound which decreases the
quantity of protein aggregate is selected.
9. A method for screening for a test compound
which increases proteasome activity for the
treatment of neurodegenerative diseases comprising
the steps of:

introducing the test compound into an animal
which models neurodegenerative disease;

allowing said animal to develop; and

- 5 subsequently measuring the quantity of
aggregates in said animal wherein a compound which
shows decreased aggregate formation over control
animals is selected.

10. Transgenic mice capable of overexpression of
HDJ-2.

Add
A2

ADDB3)

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